Society Reports.

NEW YORK NEUROLOGICAL SOCIETY.

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Frederick Peterson, M.D., President.

DOUBLE ATHETOSIS FROM INFANTILE CEREBRAL PALSY.

Dr. L. Stieglitz presented a boy, sixteen years of age, the youngest of several children. All the others had been well at birth, but he had been born asphyxiated, although the labor was normal. He did not walk until five years old. At the age of seven years he had scarlatina, and after that time the peculiar movements which he now exhibits became worse. Soon after birth it was noticed that he moved his arms, legs and head in a peculiar way. After a few years the movements remained at a standstill until the attack of scarlatina. On voluntary motion, the movements are aggravated. His face shows a constant succession of grimaces. The peculiar movements affect the trunk, as well as the upper and lower extremities. The tongue and the eyeballs are free from these movements. The deep reflexes of the arms are lively, as are also the knee-jerks, but not more than in many healthy children. His mental development is very good, but his physical development is rather deficient. Close study of the movements led Dr .Stieglitz to believe that the case was one of double athetosis, due to infantile cerebral palsy, the result of a lesion occurring at the time the child was born asphyxiated. These cases rarely show any defective mental development. In cases of athetosis the lesion is probably usually in the lenticular nucleus, or in the corpus striatum, and it is not surprising that there is no epilepsy or disturbance of the mental development.

DISCUSSION.

Dr. B. Sachs said that as he recalled similar cases that he had seen, he was forced to the conclusion that the one just presented was a most excellent illustration of the combination of the choreiform and the athetoid movements. The probability of a non-cortical origin was, of course, very great. That there was a direct relation between the typical spastic cases of infantile cerebral palsy and the choreic and choreiform athetoid cases he verily believed. In a number of cases he had found that peripheral restraint on these unruly members, kept up for months, had had a remarkaly good effect.

Dr. Edward D. Fisher said that he now had under observation a boy of the same age, who presented almost exactly the same symptoms, except that they were not quite so severe. It seemed to him that the lesion was very apt to be subcortical. The absence of epileptic conditions did not mean necessarily that the lesion was not a cortical one. The lesion was certainly not an extensive one in the motor tract.

Dr. Joseph Collins said that he agreed entirely in the diagnosis made by Dr. Stieglitz, and he was glad to see such a typical illustration of this group of cases. He did not favor the name "athetosis," nor would he apply it to this case. Taking the case in its entirety he would look upon it as an example of the choreic form of infantile cerebral palsy. He was inclined to think that the involvement was primarily of the corpus striatum and quite as much so of the optic thalamus, and he based this opinion on the study of the patient's motions, coupled with his spastic gait. The question of the existence of epilepsy in such a case did not seem to him of much importance, for the opinion was growing yearly that idiopathic epilepsy is a cortical disease. He had tried peripheral restraint for months at a time without observing the slightest benefit, even temporarily.

Dr. Peterson thought the case was undoubtedly one of post-paralytic morbid motion, and he agreed with Dr. Collins that athetosis was not a proper name. The position was athetoid, but the movement was rather a polymyoclonus. The term "athetosis" should be applied to a slow, vermicular movement.

Dr. C. A. Herter remarked that he thought the term was applied to much more rapid movements, and it did not seem to him that there was a proper pathological basis for such a distinction as that made by the last speaker.

Dr. Collins thought the term "athetosis" should be limited to rhythmical movements, and "choreic" to non-purposive, dancing movements.

Dr. Stieglitz, in closing the discussion, said that in cases of infantile cerebral palsy all the different varieties of movements mentioned were often combined, so that perhaps it would be better to select a name which would cover all of them. When the boy is less excited the movements are usually less rapid, and more vermicular in character. Under ordinary conditions, he does not present a spastic facies. The restraint treatment had been advocated by Dr. Hammond when he had first described these cases.

DISSEMINATED INSULAR SCLEROSIS.

Dr. Joseph Collins: The patient who is before you has the symptoms of two rather obscure diseases of the nervous system, and I bring him here that you may assist me in determining the one from which he suffers. He is 41 years old, married, and by occupation a silk weaver. His personal history is that he has been a user of stimulants and tobacco, the latter rather intemperately. Physically, he has led a very active life, and in many directions, particularly fencing and boxing. He has been an amateur athlete. The only point of interest in his family history is, according to his own statement, that a brother younger than him by two years, complains of the same trouble in walking that he does. When our patient was 33 years old, he had a sore on the penis for which he was treated by Dr. Geo. H. Fox. The latter assured him that this sore was of the nature of a chancroid, and gave him only local treatment. Aside from bubo, which complicated it, this terminated uneventfully in a few weeks, during which time he was treated in private practice by Dr. Fox. Aside from the attack of the grippe, which he had in 1892, he has been quite well. His present illness dates back nearly three years, the initial symptom having been, he thinks, dizziness, which was sufficiently great to interfere with his locomotion. Apparently this vertigo was not continuous, because he says that when it would come he would have to hold to something to prevent him from falling. About this time he must have had some trouble with his sight, for it appears that he went to the New York Eye and Ear Infirmary, where he was told that he had locomotor ataxia. The symptoms that next appeared after the dizziness seem to have been what he describes as piercing pains in the calves and the sensation of heavy weights in the thighs. So far as I am able to infer, this constitutes the major part of the patient's complaint until the beginning of the present summer, although it should be said that he has been sexually impotent since two years. The fact that he has been unable to work has, according to his own statement, preyed very much upon his mind and made his symptoms worse. About six mnoths ago his bowels and bladder became functionally derelict, and sometimes the one or the other would discharge their contents without his being aware of it. At this time, likewise his speech and handwriting became affected, both in very much the same way as they are now. That is, the enunciation is measured and scanning, the intonation rather high and uniform, while the handwriting is very irregular and ataxic. About the same time, the patient avers that he became uncommonly emotional. He would burst into tears at one time, and at another would grin or smile senselessly. Accompanying these physical symptoms, the patient underwent a psychical alteration. He says that he became very forgetful, that he was unable to read understandingly, even the newspapers. This state persists in a very much slighter degree to-day. For instance, it is quite impossible to elicit the symptoms of his disease except by asking him leading questions, and he forgets easily. At present, he is able to concentrate the attention sufficiently to read a newspaper or magazine understandingly.

His complaint at the present time is of difficulty in locomotion and impaired dextrality. The abnormalities of articulation and the slight psychical aberrations do not concern him sufficiently to complain of them. The functions of the bladder and bowels are now more nearly normal, although occasionally there is some incontinence, especially of urine. He does not complain of pain, nor of tight band sensation about the waist, and the feeling as if there were heavy weights in the thighs has disappeared. Examination shows a man of medium height and good muscular development, whose body is the seat of a marked deformity due to a cervico-dorsal scoliosis which he believes to have been caused by carrying heavy loads in the right hand or on the right shoulder during boyhood. As to the plausibility of such an explanation each one must determine individually. Both feet show the deformity which is known as pes cavus, pied bot of the French writers. The plantar surface of each foot is very much hollowed beneath the arch, while all the toes are forcibly extended save the first phalanx, which tends to claw the floor. When the patient stands, titubation of the whole body is the most striking phenomenon, and this persists, though to a lesser degree, when he sits. The gait is ataxic and reeling; unlike the ordinary taboid patient, he does not watch his feet while walking. He is rather more inclined to fix some point at or near his objective, and keep his vision on it until he reaches it. Station is extremely uncertain when the feet are approximated, and when he closes the eyes under such circumstances he is pretty sure to fall. The superficial, or cutaneous reflexes are present. The tendon jerks of the lower extremity are all absent, while of the upper extremity they are all present. The right palpebral fissure is somewhat larger than the left, and to explain this the patient says that it followed an attack of sore eyes. The pupils are of medium size, slightly asymmetrical, and do not respond to light. When the backgrounds of the eyes are viewed with the ophthalmoscope the left optic discs appear rather pale, a state of pink atrophy. The right optic nerve is normal. Tactile and thermal sensibilities seem to be normal. There is well marked analgesia over both legs. The patient has been examined four times only with great care, but the analgesia seemed to be present on each occasion, although varying somewhat in intensity. There is no perversion of sensibility in any other part of the body. Color sense is normal, and the visual fields are unchanged. Senses of smell, taste and hearing are intact. The trouble with speech and writing have been already sufficiently referred to above.

Now, the question arises, has this patient Friedreich's disease, or has he multiple sclerosis? Perhaps, indeed this question is not sufficiently comprehensive. Some may be inclined to ask, and legitimately, I think, can diseases of the cerebellum be eliminated on the one hand, and genuine tabes on the other? Personally, I am inclined to the opinion that the case is one of disseminated insular sclerosis, although perhaps the evidence pointing to such a diagnosis is not so direct or convincing as some might wish. If a brief analysis of the symptoms be made, it will at once be seen that there exists a certain number of conditions which are usually considered inimical to the diagnosis of Friedreich's disease. These are, first, the patient's age; second, the sensory disturbances; third, the perversion of function of the uro-genital sphere, and fourth, loss of the pupillary light reflex and paleness of the temporal side of the optic nerves. On the other hand, all of these, and the other symptoms and physical accompaniments which the patient presents, including the deformities of the trunk and foot, can be explained by the existence of islets of sclerosis in the posterior columns, in the oblongata, and possibly in the cerebellum. If my diagnosis be the correct one you will agree with me, I think, that it is one of the most atypical thus far reported.

DISCUSSION.

Dr. Sachs said he was of the opinion that the case was probably a somewhat atypical example of multiple sclerosis. There was after all not so much difference between this case of multiple sclerosis and the cases of combined sclerosis that one sees, for he had observed cases of combined sclerosis which at one stage had presented all the symptoms of a spastic paralysis, and subsequently the appearance of typical cases of tabes.

Dr. Fisher said he would agree with the diagnosis of disseminated sclerosis, basing it largely on the mental condition and the character of the speech, particularly the former.

REMARKS ON THE KIDNEYS IN CASES OF CEREBRAL HEMORRHAGE AND CEREBRAL SOFTENING.

Dr. Christian A. Herter read a paper with this title, in which he gave some of the results of his observations in 464 autopsies on adults at the Presbyterian Hospital. There were twenty-three cases of cerebral hemorrhage, eleven of cerebral softening and one of softening and hemorrhage. The three cases of hemorrhage in patients with large kidneys showed the lesions of chronic diffuse nephritis. Six of the patients with small kidneys were males, while all the patients with large kidneys were females. Of the patients with large kidneys, four had cerebral softening, two thrombosis and two embolism. The great majority of the medium kidneys were granular. In the cases of cerebral softening the kidneys presented a granular surface and freely stripping capsules. According to this series of observations, cerebral hemorrhage was most frequent among persons with small granular kidneys, and least frequent in those having large kidneys. None of the cases of hemorrhage had smooth kidneys, on the other hand, the majority of the cases of cerebral softening were associated with smooth kidneys and freely stripping capsules. Where there is a high arterial tension and the urine is of low specific gravity and contains very little albumin, and few, if any, casts, it may be inferred that the kidneys are small and granular, and this condition, in cerebral cases, points strongly to hemorrhage.

DISCUSSION.

Dr. Mary Putnam Jacobi said that she had recently seen a case which bore on the relation of uræmia and hemorrhage. The patient was a woman of about forty-five, who had been previously in excellent health. She awoke one morning to find that her vision was extremely dim. The next day she complained of double vision, and a paresis of the abducens appeared and increased until the muscle became completely paralyzed. The urine on several examinations was found entirely free from albumin, and granular casts were found without the use of the centrifuge. The urine contained a large quantity of urea. The dimness of vision was probably uræmic and the paresis of the abducens due to a very minute hemorrhage in the sixth nerve.

Dr. Herter said that it seemed to him probable that in Dr. Jacobi's case the symptoms were due to hemorrhage rather than œdema, although the latter should always be borne in mind. It was a common thing to find considerable urea during the uræmic state. This was probably to be explained by a gradual accumulation of the urea in the blood, owing to the defective action of the kidneys, and this accounted for the fact that these patients often pass a normal quantity of urea in the urine at the time that the symptoms of acute uræmia are present.

TRAUMATIC HÆMATOMYELIA, WITH REPORTS OF CASES WITH AND WITHOUT AUTOPSY.

Drs. Pearce Bailey and P. R. Bolton presented a paper on this subject from which extracts were read by Dr. Bailey. Speaking of that class of cases in which the hæmatomyelia is consequent upon injury to the spinal column, the authors said that in a large proportion the general destruction of nerve tissue is so severe that hemorrhage can have had but little influence. The blood is poured out first and most profusely into the gray matter, and this central limitation is often very sharply defined. The columnar extension of the blood is usually in the gray matter of the anterior or posterior horns, and the column extends further upward than downward. Occasionally it may extend into the white matter just behind the middle commissure. These hemorrhages are most commonly found at autopsy between the fifth cervical and the third dorsal segments. The lower dorsal or lumbar cases either do not come to autopsy or else only after so long a time that it is difficult to draw definite conclusions regarding the pathological findings.

The evolution of the lesion in spinal hemorrhage is somewhat obscure. The natural tendency is to absorption of the blood so that cavity formation is an early consequence of hæmatomyelia. A symptom of some importance is a band of thermo-anæsthesia extending a little distance above the general anæsthesia. In primary hæmatomyelia bleeding into the substance of the cord constitutes the primary lesion. There are two forms, viz.: (1) the localized; and (2) the disseminated. A force sufficient to rupture the blood vessels of the spinal cord must affect the nerve elements, hence when bleeding is prominent it is reasonable to expect lesions of the nerve cells. In the majority of cases, however, the hemorrhage is the most important factor. The focal form of primary hæmatomyelia is always found in the lower cervical or upper dorsal region. Most of the cases result from falls. One form of injury which is especially fertile in producing this injury is a sudden bending of the neck, such as occurs in falling of heavy weights on the neck, diving in shallow water, etc. The symptoms differ from those of crushes of the cord, in which the white and gray matter are both implicated The paralysis is at first flaccid everywhere, and the reflexes generally are either diminished or lost. An injury to any part of the spinal cord above the upper lumbar region, severe enough to cause paraplegia, will usually inhibit or abolish the knee-jerk for a time. This is eventually followed by an exaggeration. In the muscles of the lower extremities the paralysis usually changes within three weeks from the flaccid to the spastic type. Immediately after the accident the paralysis of motion is usually wide-spread and severe. Sensibility to touch is little, or not at all, modified in primary focal hæmatomyelia; there is, however, analgesia or thermo-anæsthesia, or both. In the cases reported the power to distinguish between heat and cold was lost while sensibility to pain was unaffected. Compared to the prognosis of secondary cases of injury, the outlook in primary focal hæmatomyelia is favorable. Primary disseminated hæmatomyelia occurs after severe general traumatism. In these cases capillary hemorrhages were found throughout the cerebro-spinal axis. Microscopic examination shows small masses of red blood cells lying, for the most part, in the gray matter, but also in the white matter. They are not accompanied by other evidence of destruction, and are not artefacts.

DISCUSSION.

Dr. Sachs said that there seemed to be every reason to suppose that in many cases which were classified as of secondary origin there must have been effusion, not only into the cord, but around the cord. On this account, the question arose as to whether many of the symptoms might not be due to compression of the root fibres. He had seen dissociation of sensation as one of the earliest symptoms of Pott's disease and Pott's paralysis. It need not, therefore, be a symptom of destruction of the gray matter. He recalled a series of cases that were not traumatic in origin, were rather slow of onset, and occurred in adults, and yet presented all the typical symptoms described under the head of hæmatomyelia. The question arose as to whether in the non-traumatic cases the peculiar arrangement of the blood vessels in the spinal cord might not account for some of the cases in his series. A case now under observation was instructive for the reason that the lower extremities alone were affected, and there was an association of the spastic symptoms with a dissociation of sensation.

Dr. Onuff said that he had seen the case just referred to by the last speaker. The patient stated that soon after a tiresome bicycle ride he noticed that he could not urinate easily. The next morning there was a tingling sensation in the legs, but it was not for five days that a partial paraplegia appeared. At this time, incontinence of fæces was also noticed. There was atrophy of the leg affecting chiefly the adductors of the right thigh. The sensation of pain was exaggerated, so that there was a marked drawing up of the limb when it was pricked. The difficulty of urination was followed by incontinence. The atrophy in the thigh would seem to indicate a higher lesion, while, on the other hand, the initial bladder and rectal symptoms would point to a lower lesion.

Dr. Fraenkel said that he thought the abolition of the reflexes in such cases as those described in the paper was of much diagnostic importance. Enough cases had been reported to show that even in the face of the absence of the reflexes it was possible for a communication to exist between the upper and lower portions of the cord. He regretted that the condition of the deep sensibility had not been recorded in the cases quoted.

Dr. Bailey remarked that in one case in which the reflexes were absent, the muscular sensation had returned.

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145. UN CAS DE SURDITÉ VERBALE PURE TERMINÉE PAR APHASIE SENSORIELLE, SUIVI D'AUTOPSIE (Word-Deafness Terminating in Sensory Aphasia; Autopsy). Dejerine and Sérieux (Journ. de Méd., Feb. 6, 1898).

In reporting this case the authors say that only four cases have been recorded of what Lichtheim called sub-cortical word-deafness, and what Dejerine has proposed to call pure word-deafness. In this condition the patient has lost simply the comprehension of spoken language and the power of repeating and writing to dictation.

The present case is one of the four and was reported by Sérieux in 1893. After that date the patient gradually grew worse and the speech difficulty became extended into a general sensory aphasia. The autopsy, with careful microscopic examination, revealed no sign of a focal lesion, but a general atrophy of the temporal convolutions, bilateral, and diminishing in degree from above downward. This atrophy was caused by changes strictly cortical, and involving primarily the cells, the more superficial ones most. The authors sum up, briefly, as follows:

1. This autopsy definitely settles the question of the localization of pure word-deafness, showing it to be due to a lesion exclusively cortical, consisting in this case of a chronic poliencephalitis, while in the case of Pick the lesion was both cortical and sub-cortical. This is the first case of the kind that has been shown to be due to a purely cellular lesion.
2. The case shows, as does that of Pick, that pure word-deafness is due to a bilateral, temporal lesion, located in the ordinary auditory centre.
3. Considering this localization, it appears probable that in pure word-deafness we have to do, not with a separation of the auditory centre from the centre of recognition of words, but with an impairment of the functions of the former. This opinion is corroborated by the fact that this patient, although she preserved the sense of hearing for a long time, gradually lost it.
4. The gradual and progressive transformation of pure word-deafness into a sensory aphasia is to be emphasized. For a considerable time the patient possessed intact the faculty of internal language, and it was only by degrees that the auditory centre for words became involved and that then appeared alexia, "jargonaphasia," and paragraphia.

PATRICK.